Gibney (V. P.)

A CASE OF

SCLERODERMA VEL MORPHŒA,

WITH

HEMIATROPHIA FACIALIS, ALOPECIA AREATA, AND CANITIES.

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Case.—Mary Marshall, æt. 9 years, was admitted to the Hospital for the Ruptured and Crippled, July 19, 1878, for the relief of talipes valgo-equinus of the left foot, induced by the disease indicated at the head of this report.

The case was first presented at the out-door department, July 13, 1876, when I observed a marked atrophy of the left side of the face, a deep, cicatricial-like sulcus to the left of the symphysis of the inferior maxilla, a roughening and mottling of the integument in the inferior maxillary region of the same side, a peculiar band-like condition of the skin over coccyx, with a like appearance over gluteal region, extending down the posterior surface of the thigh, leg, and outer border of the foot. The course taken by the disease was exactly in the distribution of the sciatic nerve. The skin over outer hamstrings was glistening, but could be caught between one's thumb and forefinger and raised a little from the tendon beneath. The comparative measurements were taken at that time, and will be compared later with those of more recent date.

The patient did not appear again until the 19th of July last,—an interval of two years elapsing,—and at that time a more thorough examination was made. It was now learned, first, that her family history, both paternal and maternal, was remarkably good; that the child herself had enjoyed good health during the first three years of life; in fact, that, with the exception of a pertussis, two years ago, and a rubeola, one month ago, her general health has always been very good. The present affection has not produced any constitutional disturbance, and all that can be learned which would indicate any deviation from health is that, for several years, she has moaned during sleep, as if in great pain, but never waking to cry out. One day, when about three years of age, there was observed by the mother a dirty-brownish discoloration in the left inferior maxillary region. It was about an inch in diameter, and could not be removed by washing. From this point the process extended upwards and towards the angle of the mouth, gradually enlarging in two years, as well as her mother can recollect, to its present dimension. No scabbing or ulceration has occurred on the face,

and no change of color at any time, unless under excitement, when the usual vaso-motor changes are observed. Over the left gluteal region a similar process began about the same time, and has acted in about the same manner. It grew to the size of one's hand, and by many was thought to have been a scar from a burn, so close was the resemblance. Extension took place downwards, and the mother did not observe any difference in the size of the limb until a year had elapsed. There has been no ulceration in its track, but at points where the deepest depressions are to be seen there has been a kind of dry scabbing observed. There has been no pruritus. Last year, for the first time, and again, two months ago, about a dozen small pimples appeared on the site of the lesion, disappearing spontaneously. The progress of case has been steady and gradual, not marked by pain or tenderness.

It is now two or three years ago since the gait became affected. This lameness has increased, though to-day she exhibits much power and endurance in a walk of about three miles. No regular course of treatment has been employed. Her complexion is fair, hair brownish, and she is fairly nourished. As she stands, the left foot is everted, and rests on the inner border, and as she walks a marked degree of lameness is observed.

To the left of the posterior fontanelle is a large lock of gray hair, a localized *canities*, while the scalp from which this grows presents no induration or atrophy, and is freely movable on the underlying tissue. The mother says this gray hair came first with the other signs of disease, and has existed about an equal length of time.

Over the left fossa is an area of baldness, one by one and a half inches in size, giving undue prominence to the blood-vessels here by the thinness of the skin, which is smooth, transparent, and mobile. This, so the mother says, has lasted a long while, and is called, Dr. Bulkley informed me some months later, *alopecia areata.*

In the inferior maxillary region, left side, the skin presents a slightly indurated appearance, mottled irregularly, and is freely movable. This area is bounded above by a line from the angle of the jaw to the angle of the mouth. From this point the margin extends to within a line of the symphysis, thence to thyroid, thence irregularly to angle of the jaw again. It corresponds closely with the distribution of the infra-maxillary branch of the facial nerve. Within this area and immediately to the left of the symphysis the skin is depressed into a sulcus, and hugs the bone closely, so that no movement can be made. Three lines to the left of this is a similar groove-like cicatrix, extending up to the angle of the mouth, drawing this angle down perceptibly. The incisors are very prominent, and the upper jaw overlaps the lower to the extent of half an inch. The lower half of the face, left side, affected by the *hemiatrophia facialis*, is about one-third smaller than that of right side. There are no marks of disease on the upper limbs or on the thorax.

In the umbilical region, to the right of the median line, is an

obliquely-oval patch of altered skin, five inches by one and seven-eighths in size. The color is pale, and the appearance in general exactly like that of an old vaccination scar without mottling or scaliness.

The skin is here freely movable, and on grasping it between the thumb and fingers it feels thinner than normal skin, although in passing the finger over the abdomen one cannot make out the boundaries of this placque; in fact, no difference can be felt between this and the surrounding skin. This has been tested by several prominent dermatologists, and they fail to recognize a difference in sensation as they pass the hand from sound to unsound skin. The mother says this has existed three or four years, though she does not know when it first appeared, and has not recognized any change from its present appearance save a disposition at times to scale.

The left buttock is flattened, and the skin (even to the right side, one inch beyond the coccyx) has a dusky-red appearance, the veins are quite prominent, and here and there we find the same vaccination-like placques, resembling the one on the abdomen, while over the trochanter and the coccyx the mobility is diminished. This hardened and bound-down condition of skin extends along the posterior surface of the left thigh, through the popliteal space, along the posterior surface of the leg, embracing both malleoli, and shading off into sound integument about the middle of the outer border of the foot. The front of the limb presents nothing abnormal. The subcutaneous cellular tissue along this tract is atrophic, and the skin lies directly upon the muscles, though not closely adherent thereto. The outer hamstring is tense and stands out prominently, while between this and the vastus externus is a groove which does not disappear when the leg is flexed. The limb can be completely extended. The group of muscles on the outer aspect of the leg are quite tense, as well as the skin overlying, and the foot is drawn in perfect valgus with moderate equinus. If the attempt is made to force the foot into a normal position, a crackling sensation is imparted to the hand placed over the tissues on the outer side. Voluntary flexion and extension are easily performed, though inversion is impossible.

Heart and lungs examined with negative results.

Comparative Measurements.

	July 13, 1876.	Feb. 25, 1879.
Thigh, right side	12 in.	14½ in.
" left side	11 "	12 "
Calf, right	8½ "	10 "
" left	8 "	7¾ "
Ankle, over malleoli, right	6 "	7 "
" " left	5¾ "	7 "
Foot in length, right	7⅜ "	8 "
" " left	7 "	7½ "

The limbs were of equal length at both dates.

The circumference of the knees, July 19, 1878, was, right, 10¼

inches; left, 9½ inches, and on Feb. 25, 1879, eight months later, right, 10¾ inches; left, 10 inches. No difference at the two dates, comparatively.

It will be seen, then, that the right thigh, the one not affected, has increased 2¼ inches in circumference within two years and eight months, while the left has increased only one inch during the same period. This makes 2¼ inches difference between the two now against 1 inch two years and eight months ago. The calves gave *then* only ½ inch difference; *now*, 2¼ inches. The right, sound one, has *increased* 1½ inch, while the left has *lost* ¼ inch in circumference.

The ankle over the bony structure, in the period named, shows no appreciable atrophy. The right has gained in the period named 1 inch, while the left has gained 1¼ inches.

The feet, as to length, show steady and equal growth, with ⅜ inch difference in length between them, and ½ inch now.

The knees in circumference eight months ago gave a difference of ¾ of an inch, and give now the same: each has increased equally.

On Nov. 30, 1878, Dr. Bulkley brought Dr. Duhring, of Philadelphia, to the hospital, and both examined the case very critically. Dr. Duhring observed an hypertrophy of the venous capillaries on both sides of the body, though especially marked in the track of the lesion.

He regarded the patch on abdomen as typical of morphœa in process of subinvolution, and felt confident that at some previous time the patch was hard. The lesion along the thigh he did not regard as true scleroderma, but rather as a condition representing the border line between scleroderma and morphœa.

The case has been presented at a meeting of the New York Dermatological Society, and is now under discussion, which discussion will appear in the Transactions of the same.

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CASE OF MORPHŒA.

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Alice Dowling, age 35, native of Ireland, unmarried, presented herself at my class at New York Dispensary in July, 1878. She is a dark-complexioned, rather stout, robust-looking woman, and her general health has always been excellent. About twelve months previous she noticed a white patch, like a scar, on the anterior aspect of her right arm, just below the flexure of the elbow. It had occasioned no pain or inconvenience, and her attention was attracted to it by accident. Two or three months later she noticed a similar patch, but larger, on upper part of same arm, and still later, a change in the color of the skin over the right cheek.

Upon examination there is found on the outer anterior aspect of forearm, just below the flexure of the elbow, a yellowish-white, oval-shaped patch of integument, distinctly circumscribed with a brownish margin. It is about one and one-quarter inches by three-quarters of an inch in size, not elevated above the surrounding skin, into which it seems to be artificially set. It has a hard, horny feel, and cannot be pinched up from the underlying tissues.

On the arm, three inches above this, is a similar patch about the size of a silver quarter.

The principal patch of the disease is situated over the region of the deltoid insertion. It is irregularly circular in shape, and measures one and one-half inches in a line corresponding to the long axis of limb, by two inches transversely. The skin has a white, waxy appearance, exactly comparable to the surface of leaf lard, and is marked by minute transverse corrugations, which give it a glazed and shiny aspect. It is perceptibly elevated, and the integument feels thickened, hard, and unyielding. On its axillary side the patch has a faint pinkish margin. Its upper and outer borders are marked by a broad margin of a mottled brownish pigmentation, interspersed with numerous milky-white spots. The lower border fades gradually into the sound skin. When the surface is scratched it yields fine laminated scales, which, when separated, have a micaceous gleam.

Scattered around this central patch, and extending two or three inches beyond its periphery, especially upwards and outwards, can be seen numerous small leucodermic spots, which are evidently outposts of the advancing disease. These white spots are each sur-

rounded by a pigmented ring, the brownish tint of which throws them in strong relief.

The area of this affected surface, taking in that invaded by the white spots, measures four or five inches in diameter. These spots can be traced down the front of the arm to the elbow, not continuously, but sprinkled here and there with areas of healthy skin intervening. They are especially numerous around an old vaccination cicatrix, the upper part of which seems to be undergoing a modification due to the disease.

Upon the right side of the face there is a circumscribed surface which appears to be undergoing the first changes of the disease, and which might be mistaken for a vitiligo. This surface is elliptical in shape; its transverse diameter, corresponding to a line drawn from the malar prominence to the angle of the jaw, measures three and one-quarter inches; its conjugate diameter, extending from the angle of the mouth to middle of the cheek, measures one and three-quarter inches. This patch is neatly defined by a pigmented margin of yellowish brown. The skin is thin, white, and delicate, presenting a faint roseate hue, in marked contrast to the freckled, brownish coloration of the surrounding skin. The hairs of the affected portion appear to have been bleached by the morbid process.

No other portions of the body are as yet involved. There is an intense itching around the patches on the arm, which provokes almost constant scratching. With this exception, there is an entire absence of all subjective symptoms.

The temperature of the principal patch, carefully tested with a Stewart's clinical thermometer, was found to be raised .8 of a degree above that of the adjacent normal skin, as well as that of the corresponding portion of the left arm.

The changes which the disease has undergone during the four months the patient has been under my observation have not been material. The patch on the forearm has grown smaller, it has lost its hard and horny feel at the periphery, and the ivory color has deepened into a dirty white, like chamois-skin.

The dead white of the upper patch is gradually merging into a yellow or yellowish-white color.